SUBCLAVIAN DOUBLE STEAL SYNDROME PRESENTING WITH COGNITIVE IMPAIRMENT AND DIZZINESS

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ABSTRACT

Background. Atherosclerotic occlusion of the brachiocephalic artery is a very rare condition resulting in a distinctive pattern of hemodynamic disturbances in the cerebral circulation described as subclavian double steal syndrome.

Case report. We present the case of a 66-year-old male patient with atherosclerotic occlusion of the brachiocephalic artery with double steal syndrome presenting with cognitive impairment and dizziness. The diagnosis was based on duplex ultrasound examination and was confirmed with angiography. The patient was referred for revascularisation surgery, with extra-anatomical bypass. The outcome was favourable regarding the vertebrobasilar symptomatology, but mild cognitive impairment persisted.

Keywords: subclavian double steal syndrome, brachiocephalic artery occlusion, cognitive impairment, dizziness

INTRODUCTION

Atherosclerotic occlusive disease of the brachiocephalic artery (innominate artery) is an infrequent condition compared with carotid or subclavian stenosis or occlusion, comprising less than 2% of all extracranial causes of cerebrovascular insufficiency. (1)

Brachiocephalic artery occlusion proximal to the origin of the common carotid and vertebral arteries may result in retrograde blood flow in one or both of these vessels, a phenomenon described as subclavian-carotid double steal syndrome. Clinical presentation varies from asymptomatic stenosis to transient or permanent cerebral ischaemia and ischaemic arm symptoms. (2,3) The atherosclerotic brachiocephalic artery occlusion develops progressively, allowing the genesis of complex collateral circulation for both the right cerebral hemisphere and right upper limb supply, which is the reason why it is usually asymptomatic. (2,3) We present a case of brachioceohalic artery occlusion where the first clinical presentation was cognitive impairment and nonspecific dizziness, followed by transient vertebrobasilar symptomatology. Written informed consent was obtained from the patient for publication.

CASE REPORT

A 66-year-old male patient with a negative medical history and with vascular risk factors (heavy smoker and alcohol consumer) presented with cog-

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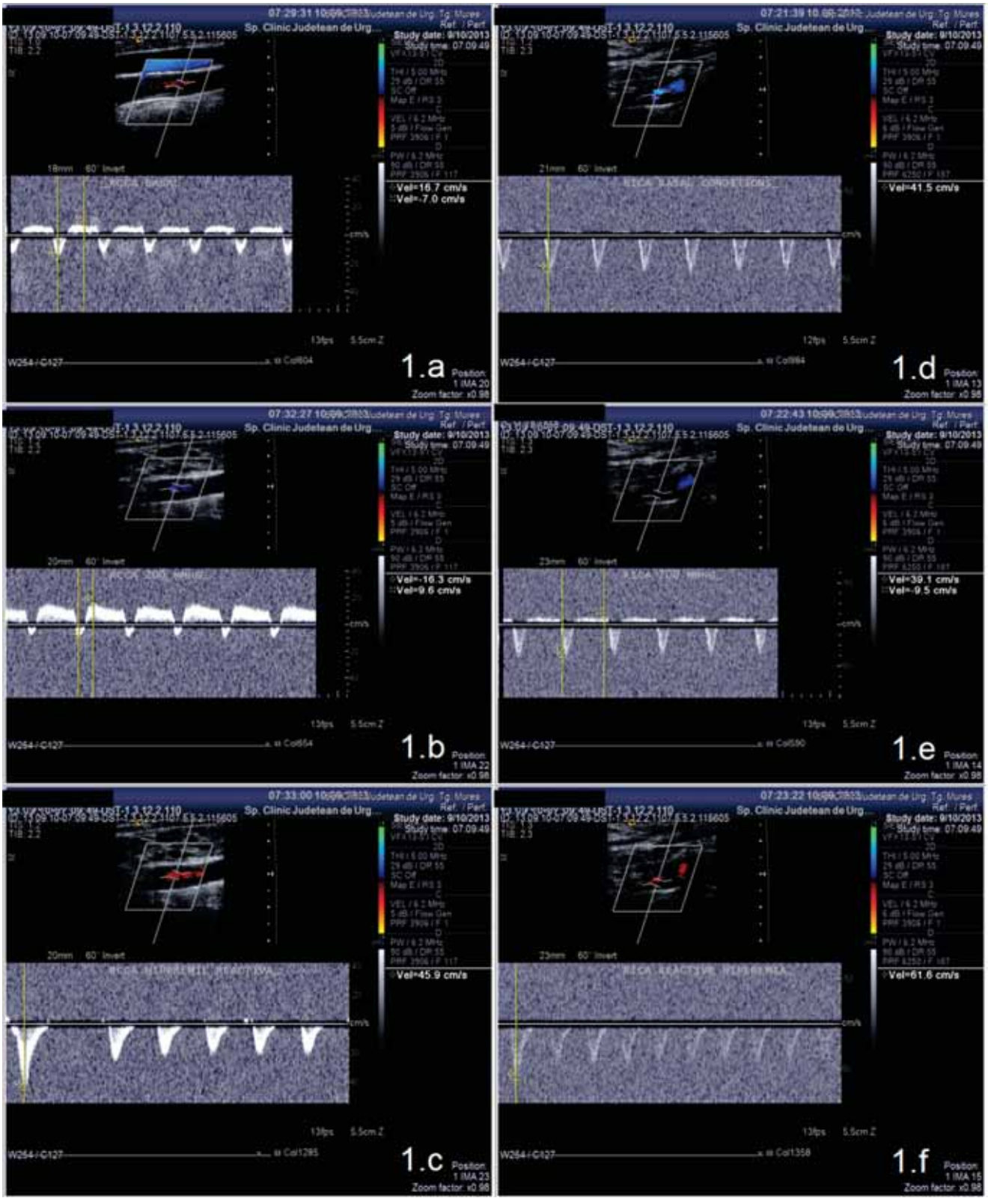

FIGURE 1. *Duplex ultrasound examination of the right common and internal carotid arteries. 1. a – Midsystolic deceleration and reversed flow in the right common carotid artery; 1. b – Decreased midsystolic deceleration, decreased reversed flow, and increased diastolic blood flow velocities in the right common carotid artery during compression of the right brachial artery with a sphygmomanometer cuff (> systolic blood pressure). 1. c – Completely reversed flow in the right common carotid artery (RCCA) during reactive hyperaemia of the right arm after cuff release. 1. d – Completely reversed flow in the right internal carotid artery (ICA); 1. e – Decreased midsystolic reversed flow and appearance of diastolic anterograde flow with low velocities in the right ICA during compression of the right brachial artery with a sphygmomanometer cuff (> systolic blood pressure); 1.f – Completely reversed flow with increased velocities during reactive hyperaemia of the right arm*

nitive impairment and dizziness lasting for 2 weeks, which worsened on the admission day and was accompanied by impaired balance. His physical examination was remarkable for the 30-40 mmHg difference in blood pressure between the two arms (right < left) and barely discernable radial, ulnar, and brachial arterial pulses on the right upper limb. Otherwise, the physical examination was unremarkable. The neurological examination at admission revealed: bilateral horizontal and vertical nystagmus; ataxic gait; diminished deep tendon reflexes in the lower limbs; mild cognitive deterioration; temporal and spatial disorientation; and Mini-Mental State Examination (MMSE) score of 26. One hour later, his neurological examination was normal exept for the cognitive impairment.

The laboratory examinations (complete blood count, blood glucose, lipid panel, urea, creatinine, sodium, potassium, and erythrocyte sedimentation rate (ESR)) were normal except the liver enzymes (aspartate aminotransferase (AST) and alanine aminotransferase (ALT)), which were slightly elevated.

Brain computed tomography and magnetic resonance imaging were performed, which showed only diffuse bihemispheric cortical atrophy. Duplex ultrasound (DU) examination of the cervical vessels revealed inverted flow in the right vertebral and internal carotid arteries and midsystolic deceleration and reversed flow in the right common carotid artery. The steal phenomenon increased after provocative manoeuvres in all three vessels (reactive hyperaemia of the arm secondary to ischaemia induced by the sphygmomanometer cuff) (Fig. 1-3).

Angiography of the cervical vessels showed brachiocephalic artery occlusion and subclavian steal (the contrast agent injected in the left subclavian artery appeared in the right vertebral artery) (Fig. 4).

The patient was referred to a vascular surgeon and was transferred to a vascular surgery clinic for revascularisation. They performed bypass surgery between the ascendent aorta and bifurcation of the brachiocephalic artery with a Dacron prosthesis. The post-surgical evolution was favourable, the dizziness disappeared, the neurological examination was normal except for mild cognitive impairment.

DISCUSSION

The steal phenomenon occurs when the distal segment of an obstructed artery is perfused through collaterals that divert blood from adjacent arterial territories. They develop whenever there are gradients from high to low pressure systems and adequate arterial anastomoses (4). The term "steal syndrome" is used when diversion of flow is accompanied by symptoms of deficient circulation and "steal phenomenon" or "steal effect" when the steal is asymptomatic. (4)

Reversed flow in the vertebral artery can be caused by subclavian artery, brachiocephalic artery, or aortic arch obstruction. Reversed flow in the carotid artery can be caused by brachiocephalic artery occlusion. (4)

The left subclavian artery is involved in subclavian steal three times more frequently than the

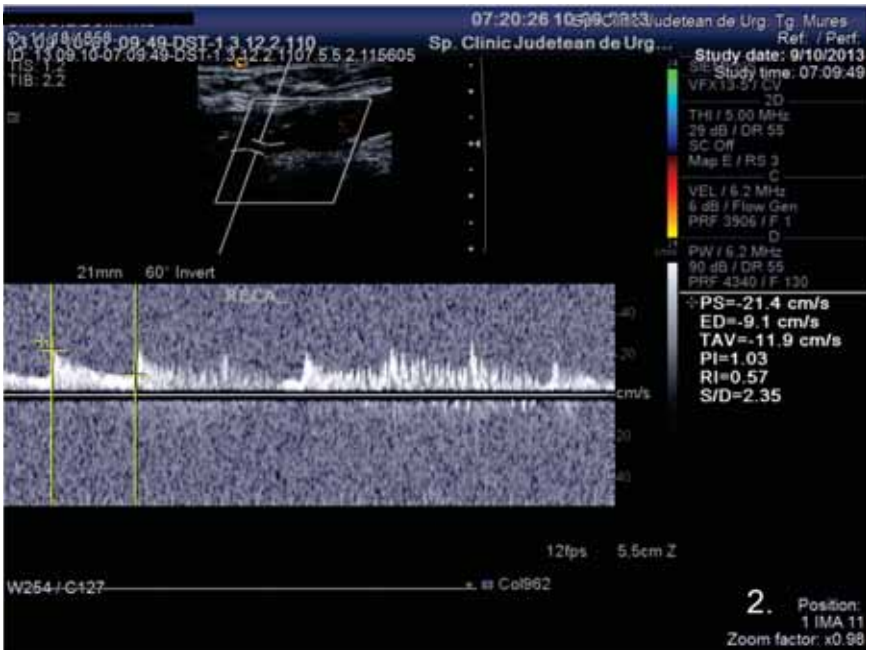

FIGURE 2. *Duplex ultrasound examination of the right external carotid artery (ECA). Anterograde flow with a poststenotic pattern*

right. Fields and Lemak (5) found in the Joint Study of Extracranial Arterial Occlusion that 17% of 6534 patients had arteriographic evidence of subclavian and/or innominate stenosis of greater than 30% or occlusion. However, only 168 patients had symptoms of subclavian steal syndrome.

Occlusive and severe stenotic diseases of the brachiocephalic artery are relatively rare, account-

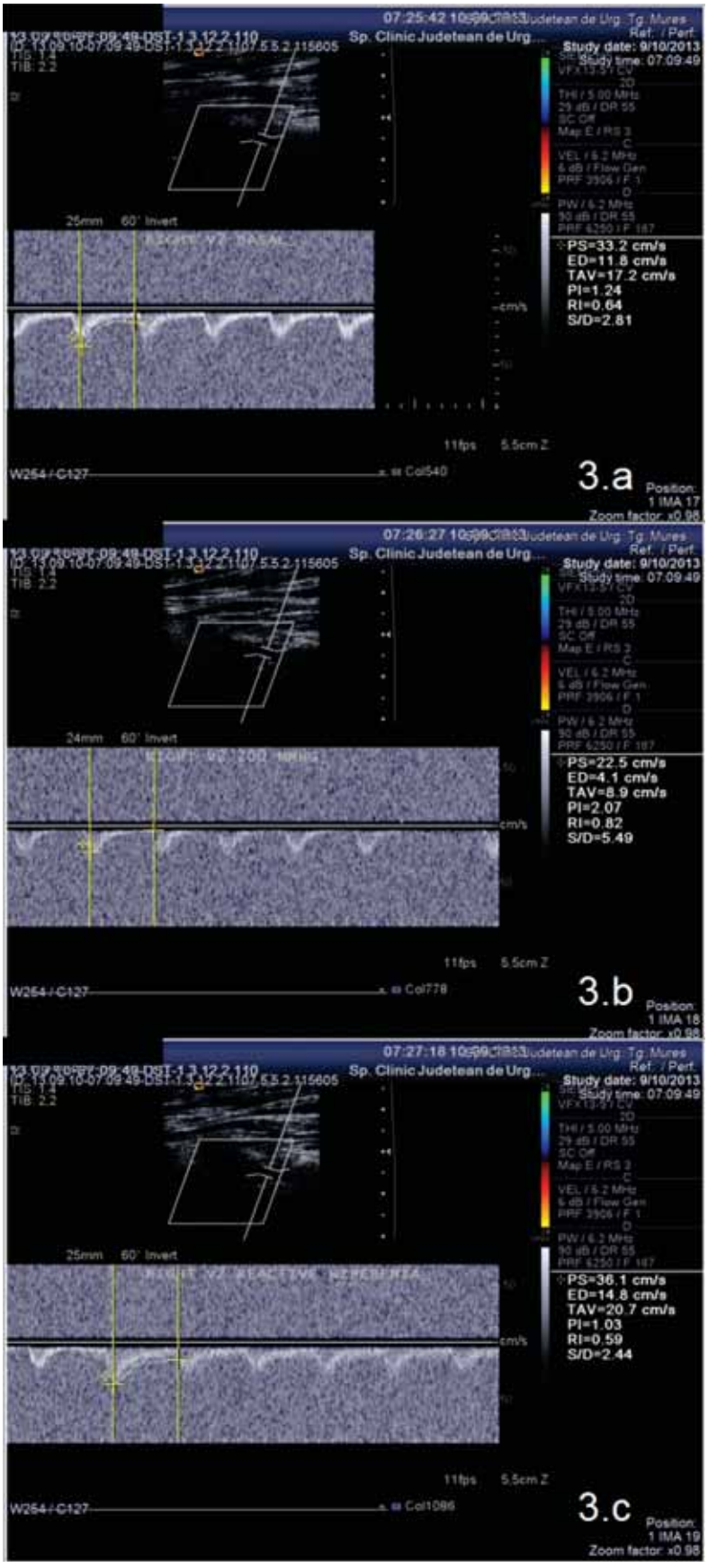

FIGURE 3. *Duplex ultrasound examination of the right vertebral aretry (V2 segment). 3.a - Completely reversed flow during basal conditions; 3.b - Completely reversed flow with decreased flow velocities during compression of the right brachial artery with a sphygmomanometer cuff (> systolic blood pressure); 3.c - Completely reversed flow with increased flow velocities during reactive hyperaemia of the arm after cuff release.*

FIGURE 4. *Digital subtraction angiography (DSA) demonstrating the subclavian steal phenomenon. The contrast agent injected in the left subclavian artery near the origin of the vertebral artery appeared in the right vertebral artery.*

ing for as much as 2.5-4% of atherosclerotic lesions of the extracranial and intracranial cerebral arteries. Brunhölzl and von Reutern (6,7,8) identified in their study only 20 cases of haemodynamically significant lesions of brachiocephalic artery among 30,000 patients.

In our database of 7000 cervicoverebral DU records performed over a 6-year period for patients with acute and chronic cerebrovascular pathology (2008–2013, Mures County Clinical Emergency Hospital, Targu-Mures, Romania), we found only one case (the presented case) with brachiocephalic occlusion (0.01%).

The steal phenomenon secondary to innominate artery occlusion is more frequently symptomatic compared with steal secondary to subclavian stenosis or occlusion. In the case the series reported by Grant et al., (2) eight of 12 patients presented a wide veriety of symtoms related to both anterior circulation (amaurosis fugax or carotid territory stroke) and posterior circulation (syncope and cerebellar infarction). In the case series reported by Brunhölzl and von Reutern, (7) 10 of 20 patients presented vertebrobasilar symptomatology, 7 were asymptomatic, and 3 presented carotid territory symptoms (transient ischaemic attack or infarction). In the case of our patient, the symptomatology was related to both anterior (cognitive symptoms) and posterior circulation (dizziness, nystagmus, and dysequilibrum).

The following collateral pathways can divert blood to the distal subclavian artery in case of proximal subclavian artery or brachiocephalic artery occlusion: the controlateral vertebral artery; internal carotid artery through the posterior communicant arteries; the occipital branch of the ipsilateral external carotid artery; the muscular branches of the vertebral arteries; the costocervical or thyrocervical trunks of the thyroid axis; and the internal mammary and intercostal arteries. (4, 9)

DU is a useful, noninvasive method for examination of the cervical vessels and is currently the initial examination for most of the patients with cerebrovascular disease. DU can reveal the haemodynamic changes secondary to severe subclavian and innominate artery pathology. The waveform changes in case of brachiocephalic occlusion are similar to those described in subclavian steal syndrome, where the reversed flow of the ipsilateral vertebral artery is the most important component. In the case the series reported by Grant et al., (2) there was complete reversal of vertebral artery flow in eight of 12 patients at rest, and the carotid circulation was affected in fewer cases. Doppler findings in the carotid system ranged from mild midsystolic deceleration to complete reversal of flow; the latter was present only in one of 12 patients. The midsystolic deceleration and flow reversal are less frequent in the external carotid arterythan in the common carotid artery and internal carotid artery,

probably due to the rich collateral bed between the right and left external carotid arteries. (2) In our case, the flow was completely reversed in the right vertebral and internal carotid arteries, and there was midsystolic deceleration and reversed flow in the right common carotid artery and anterograde flow in the right external carotid artery. Because of the small number of cases, the sensitivity and specificity of ultrasonography for the diagnosis of brachiocephalic occlusion is unknown. (2)

According to the literature, surgical intervention is indicated in symptomatic cases, and the goal of intervention is the restoration of permanent and stable anterograde flow in the extracranial vessels, correcting the cerebral hypoperfusion and its associated symptoms (3). Extra-anatomical bypass surgery is frequently used. (3) We did not find data in the literature regarding cognitive decline in subclavian double steal syndrome and its outcome after surgical treatment.

In conclusion, we present a rare case of subclavian double steal syndrome with an unusual clinical manifestation (cognitive decline and dizziness). The diagnosis was based on DU examination, which revealed the distinctive haemodynamic alterations in the right carotid and vertebral arteries. The vertebrobasilar symptomatology was completely reversed after revascularisation surgery, but the cognitive impairment was persistent.

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